Introduction

Bronchiectasis is a disorder that has long been considered to be a diagnosis in itself but a pathological description of abnormal irreversible airway morphology. The condition is characterised by dilated airway calibre and thickened bronchial wall. Bronchiectasis not caused by cystic fibrosis (NCFB) is perceived to be rare in developed countries. The fall in prevalence being attributed to improved socio-economic conditions, vaccinations and ease of access to antibiotics. But NCFB remains an important cause of respiratory morbidity in the developing world. Chronic infection is thought to play a pivotal role in the development of bronchiectasis; however, the condition is widely believed to complicate underlying defects of host defence or pulmonary anaatomic defects.

A vicious cycle of impaired mucociliary clearance, infection and inflammation has been proposed as the most likely mechanism by which irreversible airway damage occurs. Plain chest radiographs are not sensitive enough to diagnose bronchiectasis, and when compared to High resolution thoracic CT (HRCT) there is agreement in only 5% of cases. In a paediatric series, HRCT is considered the gold standard method for the diagnosis of NCFB. The principal characteristics of the lesion on HRCT include: a) axial section diameter of or more bronchomegaly greater than that of adjacent pulmonary artery b) mucoid impaction within a dilated bronchus c) non-tapering bronch in cuts parallel to the direction of travel of bronchial lumen and d) bronchi visible adjacent to non-medial pleura.

Nissan’s Fundoplication (2). Eighteen children with asthma were at BTS guidelines for the period 1996-2006. Cases were verified following chart review, exclusion of CF and radiology review of each High resolution thoracic CT. Cystic Fibrosis was excluded by a negative sweat test (sweat chloride <60 mMol/L) by quantitative plicofacile airway clearance (and/or genotype with two mutations plus clinical sight and symptoms). Specific diagnostic evaluation was determined by each individual's clinical presentation. However the following investigations: (sweat test, sputum microbiology, differential white cell count, immunoglobulins, complement levels and specific antibody response to pneumococcus, haemophilus and tetanus) were performed in all children while extended immunologic evaluation, genetics, Mantoux test, lower oesophageal pH probe, pulmonary function tests, barium swallow, video fluorescence bronchoscopy, bronchial saline, ciliary beat frequency under light microscopy and nasal & bronchial brushings for ciliary ultra structure under electron microscopy were performed where clinically indicated.

Results

Ninety five (95) patients met the criteria for the study and were included. The median age at diagnosis was 6.4 years (range 1.5-13 years). Chronic wet cough, wheeze and recurrent respiratory tract infection were the commonest presenting complaints. Median age at onset of symptoms was 3.9 years (range 1-12 years). An underlying aetiology was determined in 86 (90%) of patients (Table 1). The commonest cause n=16 (17%), followed by immune deficiency n= 15 (16%) and chronic aspiration n= 15 (16%). Primary ciliary dyskinesia accounted for 8 (8%) (Table 1).

Discussion

Over time patients were discharged each year with a new diagnosis of NCFB. This study reflects the collective in patient experience of the condition in Dublin's three childrens hospitals (DCH) over a decade and is probably a good surrogate for the situation in the Republic of Ireland. DCHs care for most of the country's tertiary respiratory paediatric referrals and one of the centres is the designated national centre for paediatric cardiothoracic and complex malformation surgery and hence it is assumed that all cases of NCFB requiring tertiary evaluation would have been captured in this study. However, HIPE data only reflects discharges and by definition the method would miss patients diagnosed as outpatients and those older children and adolescents in the care of chest physiotherapists cared for in adult hospitals. Given these limitations, it would not valid to make any inferences regarding incidence & prevalence. Persisting high rates of childhood NCFB have been reported in Pacific Islands and Native Alaskans. Australian aborigines have rates as high as 14.7 per 1000 children. In contrast, Finland has incidence of 0.5 per 100,000 children, diagnosed using HRCT. In Irish children, diagnosed using HRCT. We report the experience of NCFB in the three Dublin Children's Hospitals from 1996-2006.

Methods

The hospital in-patient enquiry system (HIPE) is a soft ware programme used in the Irish health system to record clinical data and diagnosis for all discharges from the Irish public hospital service. The computerized system was used to identify children (<18 years) with chronic bronchitis, bronchiectasis and chronic suppurrative lung disease. In order to identify cases of NCFB in the three Dublin Children's Hospitals for the period 1996-2006. Cases were verified following chart review, exclusion of CF and radiology review of each High resolution thoracic CT. Cystic Fibrosis was excluded by a negative sweat test (sweat chloride <60 mMol/L) by quantitative plicofacile airway clearance (and/or genotype with two mutations plus clinical sight and symptoms). Specific diagnostic evaluation was determined by each individual's clinical presentation. However the following investigations: (sweat test, sputum microbiology, differential white cell count, immunoglobulins, complement levels and specific antibody response to pneumococcus, haemophilus and tetanus) were performed in all children while extended immunologic evaluation, genetics, Mantoux test, lower oesophageal pH probe, pulmonary function tests, barium swallow, video fluorescence bronchoscopy, bronchial saline, ciliary beat frequency under light microscopy and nasal & bronchial brushings for ciliary ultra structure under electron microscopy were performed where clinically indicated.

The most common co morbidities were asthma, n=18 (20%) and GORD n=10 (11%). In 23 children mean FEV1 was 82% of predicted value and FVC was 84% predicted value. Fifty one (51%) children had only one lobe involved. The most frequently affected location was the left lower lobe followed by the right middle lobe (Table 2). Haemophilus influenzae was the commonest sputum pathogen (Table 3). Airway clearance was recommended for all patients. Of 92, 23 (25%) underwent surgical intervention. Lobarectomy (13), bronchobectomy (2), bronchial stent repair 4, tracheo-oesophageal fistula and 10 cases underwent transplarmonary aspiration. Nissans fundoplication (2). Eighteen children with asthma were at BTS guidelines step 2 or higher. Eight patients were receiving regular immunoglobulin replacement therapy for immune deficiency states.

Our findings in terms of jeotology, lobar distribution and sputum pathogens are similar to other studies. Approximately one third of cases will have no specific underlying diagnosis. In the future, it can be anticipated that the diagnostic yield will improve as our knowledge of immune dysfunction expands and better methods for diagnosing primary ciliary dyskinesia (PCD) and pulmonary aspiration emerge. The
diagnosis of PCD remains challenging and undiagnosed the condition remains under diagnosed. The estimated incidence of this autosomal recessive disorder is 1 in 15,000 live births. Early diagnosis and the institution of airway clearance and other specific therapies, where appropriate will prolong lung function and lead to a loss of pulmonary morbidity in adulthood. In the current series symptoms were present for almost three years before the diagnosis was made. The presence of wheezes may be incorrectly attributed to diagnosis of asthma thus leading to further delays in diagnosis. The matter is further complicated by the co-existence of asthma in 20% of the series, which is consistent with other reports. Final diagnosis may lead to frequent lung resections and permanent loss of lung function. Milder forms of NCFB may resolve in younger children with appropriate conservative management. Our experience suggests that NCFB remains a significant problem in Irish children despite improvements in the country's socio-economic status.

Correspondence: P Greally
Department of Paediatric Respiratory Medicine, National Childrens Hospital, Tallaght, Dublin 24
Email: peter.greally@amnch.ie

References

Addendum

Bronchiectasis in Children

Bronchiectasis not caused by cystic fibrosis (NCFB) is considered an orphan disease. First described by Laennec in 1819, it is not a diagnosis in itself, but rather a result of chronic inflammation that causes irreversible damage to the lung. This damage occurs as a result of chronic infection of the bronchi, which leads to irreversible abnormalities in the structure of the bronchi. It is postulated that bronchiectasis, although a clinical finding, is caused by an underlying chronic respiratory tract infection. In the absence of a specific cause, bronchiectasis is considered an idiopathic condition.

It is postulated that a vicious cycle of infection, inflammation and impaired mucociliary clearance leads to irreversible airway and parenchymal destruction. Most adult NCFB originates in childhood. It is estimated that 100,000 individuals are being treated for the condition worldwide. However, there are no published comparative epidemiologic data.

The diagnosis may be difficult and is often delayed. Common to many studies of childhood bronchiectasis is the delay in diagnosis and the confusion of symptoms with those of asthma. A chronic wet cough or recurrent wet cough are prominent symptoms and should prompt a search for asthma. A poor response to asthma therapy should also prompt a clinical suspicion of NCFB. Consider referral to a respiratory paediatrician if clinical suspicion exists.

Bronchiectasis not caused by cystic fibrosis (NCFB) is considered an orphan disease. First described by Laennec in 1819, it is not a diagnosis in itself, but rather a result of chronic inflammation that causes irreversible damage to the lung. This damage occurs as a result of chronic infection of the bronchi, which leads to irreversible abnormalities in the structure of the bronchi. It is postulated that bronchiectasis, although a clinical finding, is caused by an underlying chronic respiratory tract infection. In the absence of a specific cause, bronchiectasis is considered an idiopathic condition.

Despite advances in socio-economic conditions over the last 3-4 decades respiratory paediatricians continue to diagnose bronchiectasis with regularity. Almost 15% of all respiratory referrals to a specialist clinic in Newcastle, UK have been subsequently diagnosed with the condition.

In this issue, we report a retrospective review of the condition in the three Dublin children's hospitals over the last decade. More than 9 new cases were identified each year. The incidence of NCFB in 92 cases was similar to other published series.1.4 In a prospective evaluation of the prevalence of NCFB we have identified cases in the Republic of Ireland. The incidence of NCFB in the country's socio-economic status suggests that NCFB remains a significant problem in Irish children despite improvements in the country's socio-economic status.

P Greally, AA Zaid
Department of Paediatric Medicine, National Childrens Hospital, Tallaght, Dublin 24
Email: pete@pcrc.net

References

In conclusion, the presence of bronchiectasis in children remains a major problem in developing countries. However, in Ireland as living standards have improved its occurrence has undoubtedly reduced. However, there are no published comparative epidemiologic data.